Pulmonary Thromboembolectomy in Chronic Thromboembolic Pulmonary Hypertension: A Case Report and Review of Literature

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ABSTRACT

Chronic thromboembolic pulmonary hypertension (CTEPH) is an important cause of severe pulmonary hypertension (PH) resulting in significant morbidity and mortality. Chronic thromboembolic PH occurs when a pulmonary embolism fails to undergo complete thrombolysis leading to vascular occlusion and pulmonary hypertension. Despite the fact that CTEPH is a potential consequence of pulmonary embolism, diagnosis requires a high degree of vigilance as many patients will not have a history of thromboembolic disease. The ventilation perfusion scan is used to evaluate for the possibility of CTEPH although right heart catheterization and pulmonary artery (PA) angiogram are needed to confirm the diagnosis. Pulmonary thromboendarterectomy is the first-line treatment for patients who are surgical candidates. This case report and review describes the pathophysiology, risk factors, diagnosis, and management of CTEPH. As it is a potentially curable cause of PH, its accurate diagnosis is vital. The gold standard and effective treatment for CTEPH is pulmonary endarterectomy (PEA). Pulmonary endarterectomy is an uncommon procedure with less than 50 years of experience worldwide. Research on the development of new surgical approaches is essential. In the present case, a new successful surgical technique for PEA was introduced.

Conclusion: The surgical procedure used on the present patient was a unique technique. We do not claim that our technique is better than the original San Diego technique, but it is suggested as a modification that may improve patient survival. However, this procedure has its own limitations and cannot be used for clots that are located distally. Therefore, further experience should be obtained in order to overcome the limitations and improve the applicability of the technique.

Keywords: Chronic obstructive pulmonary disease, Chronic thromboembolic pulmonary hypertension, Interstitial lung disease, Pulmonary artery angiogram, Pulmonary endarterectomy, Pulmonary thromboendarterectomy.

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INTRODUCTION

Chronic thromboembolic pulmonary hypertension (CTEPH) is an important cause of severe pulmonary hypertension (PH) resulting in significant morbidity and mortality. As it is a potentially curable cause of PH, its accurate diagnosis is vital. The gold standard and effective treatment for CTEPH is pulmonary endarterectomy (PEA). Pulmonary endarterectomy is an uncommon procedure with less than 50 years of experience worldwide. Research on the development of new surgical approaches is essential. In the present case, a new successful surgical technique for PEA was introduced.

CASE REPORT

A 45-year-old woman presented with a chief complaint of dyspnea and fatigue. She had a history of dyspnea and exertional chest pain for 6 months, which had worsened in the last few months (New York Heart Association functional class III). She had no history of orthopedic surgery, deep vein thrombosis, being bed ridden, or long air travel. Physical examination revealed an elevated neck vein, loud P2, tachypnea, and light edema of the feet.

In evaluations performed previously in another medical center, an echocardiographic study revealed normal left ventricular systolic function and size, severe pulmonary arterial hypertention (PAH), Grade 1 tricuspid regurgitation, and dilated inferior vena cava with minimal collapsibility. Based on echocardiographic data, the estimated systolic pulmonary pressure was 130 mm Hg.

As the chest X-ray and the pulmonary function test were normal, it seemed that the patient’s PH was not related to chronic obstructive pulmonary disease (COPD) or interstitial lung disease (ILD). A lung computed tomography (CT) scan with contrast angiography
was performed. It revealed a filling defect compatible with chronic pulmonary thromboembolism in the right pulmonary artery (RPA) leading to its complete obstruction (Fig. 1). It was also decided to perform right heart catheterization to measure the patient’s hemodynamic parameters in order to confirm CTEPH.

All findings were suggestive of CTEPH, therefore, surgical embolectomy was recommended. Evaluation of the patient prior to surgery excluded predisposing factors of CTEPH including antiphospholipid antibodies, anticardiolipin antibodies, lupus anticoagulant, and inflammatory bowel disease.

**Surgical Technique**

The patient was intubated, and after inserting monitoring devices, the median sternotomy was performed. Cannulae were inserted into both the vena cava and the ascending aorta encircled with tapes. Cardiopulmonary bypass was established under moderate (26°C–31°C) hypothermia. The aorta was clamped, and a cold cardioplegia solution was infused into the aortic root with subsequent infusions of the same solution every 20 minutes. During cooling, the superior vena cava (SVC) was immobilized to the level of innominate vein, but the azygos vein was not divided. Right pulmonary artery was mobilized by retracting the vena cava laterally and the aorta medially by using encircling tapes. After establishing cardiac arrest, on the RPA, a 4 cm incision was taken. A plane was established using a sharp dissector, and thromboembolectomy was done. Establishing the correct plane is important as a very deep plane will result in perforation of the vessel and a plane that is too superficial will result in inadequate embolectomy. After completing the dissection in the distal part of the RPA, the incision on the proximal part was extended, and then, the embolus material was removed (Fig. 2). Finally, the RPA incision was repaired with Prolene 5-0.

The cardiopulmonary bypass duration was 1 hour 10 minutes, and the aortic clamp duration was 50 minutes. After re-warming and heart beat restoration, cardiopulmonary bypass was discontinued, the cannulae were removed, and the rest of the surgery was completed under standard conditions. The patient was transferred to the intensive care unit. She was hospitalized for 7 days and discharged in good general condition. The postoperative day 2 echocardiogram showed a significant decrease in the size of the right heart chambers, decreased systolic pulmonary pressure (from 130 to 60 mm Hg), improved right ventricular (RV) function, and reduced tricuspid valve regurgitation. The lung CT scan with contrast angiography showed complete reopening of the RPA and its lobar branches.

**Literature Search Methods**

The literature search of electronic databases, such as MEDLINE, EMBASE, Cochrane Library, Canadian Medical Association InfoBase, and National Guideline Clearinghouse, etc. were undertaken by way of combining heading terms and text search terms like, ‘pulmonary thromboendarterectomy’, ‘chronic thromboembolic pulmonary hypertension’, ‘chronic pulmonary embolism’, and ‘PEA in order to identify the body of published evidences on chronic thromboembolic PH.

**Literature Search Results**

Pulmonary embolism is a common condition. Some patients subsequently develop CTEPH. Many care gaps exist in the diagnosis and management of CTEPH patients including lack of awareness, incomplete diagnostic assessment, and inconsistent use of surgical and medical therapies. Asymptomatic patients postpulmonary embolism should not be screened for CTEPH. In patients with PH, the possibility of CTEPH should be routinely evaluated.
with initial ventilation/perfusion lung scanning, not CT angiography. Pulmonary endarterectomy surgery is the treatment of choice in patients with surgically accessible CTEPH, and may also be effective in CTEPH patients with disease in more ‘distal’ PAs. The anatomical extent of CTEPH for surgical PEA is best assessed by contrast pulmonary angiography, although positive CT angiography may be acceptable. Novel medications indicated for the treatment of pulmonary hypertension may be effective for selected CTEPH patients. Chronic thromboembolic PH is defined as follows:

- A mean pulmonary arterial pressure (mPAP) of 25 mm Hg or greater and pulmonary vascular resistance (PVR) of 3 Wood units (240 dyne × sec/cm²) or greater
- Persistent angiographic pulmonary arterial thrombotic obstruction despite at least 3 months of effective, uninterrupted anticoagulation

Clinical recognition and management of CTEPH are important for several reasons. First, CTEPH is believed to be one of the most common causes of PH. Second, CTEPH is a serious, progressive and often fatal disease. Patients with untreated CTEPH experience significantly increased mortality—observational studies have estimated the median survival rate in severe CTEPH patients to be as low as 10 to 20% at 2 to 3 years. Third, CTEPH is potentially curable with PEA surgery. Finally, CTEPH patients may also benefit from treatment with novel PH-specific medications that are currently available for patients with other types of PH such as PAH.

DISCUSSION

Chronic thromboembolic pulmonary hypertension is a disabling disease referred to a late onset complication of pulmonary thromboembolism that decreases the patient’s functional status and reduces the patient’s chance of survival. The main cause of death in such patients is usually RV failure. Chronic thromboembolic pulmonary hypertension is misdiagnosed in many cases as occurred for our patient at his first visit. The patient’s dyspnea had been previously attributed to COPD or ILD. As no positive findings were obtained by chest X-ray and pulmonary function test (PFT), performing the lung CT scan with contrast angiography indicated a filling defect compatible with chronic pulmonary thromboembolism in the RPA leading to its complete obstruction.

The available treatments for CTEPH include medical therapy, PEA, and pulmonary transplantation. Lung transplantation is not recommended as the first step due to its unsatisfactory results: a postoperative mortality rate of 20% and a 5-year survival rate of 50%. Medical therapy is appropriate only in patients with inoperable or residual postoperative CTEPH. The treatment of choice for CTEPH is surgical PEA leading to normal pulmonary hemodynamics in 80% of the patients. The pathophysiology of PH and RV overload in many CTEPH patients is related to the presence chronic, organized thrombotic vascular disease at the level of the larger proximal PAs including the main, lobar and segmental PAs. By definition, such disease is not treatable with simple anticoagulation because the occlusive material is believed to have evolved from a thrombus to more organized or fibrotic tissue. No medications have been shown to be effective in treating this occlusive pulmonary arterial disease.

The current surgical procedure for PEA as a standard approach to the treatment of CTEPH was first developed in the late 1980s at the University of California at San Diego (California, USA) by the team led by Dr Ken Moser. Since the initial reports, there have been several modifications to the surgical approach to PEA. Approximately 4000 procedures have been completed in specialized centers in many countries worldwide.

The accepted approach to the PAs is through a median sternotomy using central cannulation with cardiopulmonary bypass. Due to bronchial artery hyperplasia in CTEPH, PEA is usually performed under deep hypothermic circulatory arrest or low-flow bypass to minimize bleeding, and to optimize visualization and the quality of the PA dissection. After aortic cross-clamping and the administration of myocardial protection with cardioplegia, the right and left main PAs are sequentially approached through arteriotomies extending out close to the pericardial reflection. The RPA is approached from the space between the SVC and the aorta. Once the blood vessel is opened, an appropriate endarterectomy plane is developed in the posterior wall of the vessel using blunt dissection. The specimen is prepared circumferentially, with subsequent careful dissection distally into the lobar and segmental vessels of each lung. The periods of circulatory arrest are generally limited to a maximum of 20 minutes, with corporeal reperfusion for 10 minutes between periods of arrest. Two 20 minutes period are usually required for complete excision of bilateral specimens. After the specimen has been completely removed, the PA is closed, and the patient is subsequently rewarmed and weaned from cardiopulmonary bypass. Postoperative care is usually in an intensive care unit setting with routine clinical and hemodynamic monitoring.

Most probably, the first PEA was performed in May 1962 by Dr Charles Hufnagel. In 1970, Nina Braunwald performed the first operation using a right lateral thoracotomy and cardiopulmonary bypass at the University of California San Diego (UCSD). Since
then, the technique has been modified progressively, including the use of median sternotomy and hypothermic circulatory arrest, more proximal incisions, an approach to the right side beneath the SVC rather than above it, and the avoidance of more than one arteriotomy on each side. The technique for endarterectomy was mainly developed by Dr W Jamieson at UCSD. In the routine procedure, after circulatory arrest is established, an incision is made in the PA between the aorta and the SVC. Any loose thrombotic material debris, if present, is removed. The correct plane is established with a sharp dissector, and intima and a part of media are removed. Then, the fibrotic material is grasped gently with a pair of forceps while sweeping away the outer vessel wall layer with an aspirating dissector resulting in the progressive withdrawal of the endarterectomy specimen.15

Although PEA is the gold standard of treatment, its perioperative mortality is about 10% (4–24%).16–20 Yet, PEA remains an unusual procedure. The method does not support complete clot removal, and the outcome is highly dependent on the degree of thrombotic specimen extraction and damage to the PA bed. The aspirating dissector can be harmful for the PA and may cause perforation in the PA. Pulmonary endarterectomy is the best treatment for patients with CTEPH. Traditionally, PEA has been performed utilizing deep hypothermic circulatory arrest to provide a bloodless field, but some recent reports have challenged this concept. We reviewed our experience with selective extra suctions as the initial strategy of controlling bronchial collateral flow to avoid complete circulatory arrest in patients undergoing PEA.

In our center, PEA was done as mentioned above with the exception that extra suctions were used instead of an aspirating dissector. The modification seems to be safe and convenient. In conclusion, the surgical procedure used on the present patient was a unique technique. We do not claim that our technique is better than the original San Diego technique, but it is suggested as a modification that may improve patient survival. However, this procedure has its own limitations and cannot be used for clots that are located distally. Therefore, further experience should be obtained in order to overcome the limitations and improve the applicability of the technique.

CONCLUSION

Pulmonary endarterectomy has become the standard of care given the dramatic hemodynamic and clinical improvements that have been observed in many CTEPH patients. This is in contrast to the poor prognosis for survival that has historically been associated with CTEPH.1,4 For example, survival at 2 years was less than 20% when mPAP exceeded 50 mm Hg in non-surgically treated CTEPH patients in one study from an era before the availability of PH-specific medications.2 A significant mortality of 32% was also found in CTEPH patients with less severe hemodynamics.3

Key Evidence

Although approximately 4000 PEA surgeries have been performed worldwide in the past 30 years, not all of these patients’ outcomes are reported in the published literature. A large number of observational reports have described the effects of PEA surgery on pulmonary hemodynamics,21 cardiac size and function,22,23 clinical parameters and other important outcomes, such as survival.9,24,25 However, there are no RCTs that directly compared PEA surgery for patients with surgically accessible CTEPH with either conservative management alone (e.g. diuretics, oxygen and anticoagulation) or with conservative management plus novel PH-specific medications without PEA.

In the vast majority of reports, there is an immediate improvement in hemodynamics after PEA including an increase in CI and significant decreases in PVR and PAP both on arrival to the intensive care unit and over the ensuing few days.9 Right ventricular remodeling also occurs quite rapidly after PEA.26,27 There is an immediate decrease in right-sided chamber sizes, a marked reduction in tricuspid insufficiency, with normalization of valve geometry, decreased leftward shift of the interventricular septum, increased left ventricle (LV) end-diastolic area and reduced LV eccentricity.28–30 Clinical improvement after PEA is likely related to improvement in pulmonary hemodynamics,31,32 blood flow and RV function following removal of pulmonary arterial obstructive material29,33 but may also be due to reversal of pulmonary vascular remodeling.34,35

In experienced hands, perioperative (30-day) mortality ranges from 4 to 10%, with the most common cause of early death related to persistent PH.24,25,29,36 In the largest series,36 PEA perioperative mortality between 1998 and 2002 was 4.4% (22 of 500). Increasing surgical experience, technical refinements and better patient selection likely explain the improvements in perioperative mortality during the past 15 years. However, the purported benefits of PEA suffer from potential publication bias, and outcomes37–39 may vary significantly from center to center depending on experience and surgical expertise.

Among survivors of PEA, there is evidence that patients can expect significant improvement in long-term outcome. A comprehensive follow-up conducted in San Diego (California, USA) between 1970 and 1994 reported a survival rate of 75% 6 years after surgery in 514 PEA patients. The most common cause of late death
was residual PH post-PEA, with death in this subgroup occurring at a mean of 2.73 years after surgery. More recent series have found 2-year survival to be in the 85 to 90% range\textsuperscript{24,40-42} in striking contrast to the natural history studies of nonsurgically treated CTEPH patients.\textsuperscript{1}

Studies have also reported significant post-PEA improvements in long-term health-related quality of life (HRQoL),\textsuperscript{40,41,46} exercise tolerance as measured by six-minute walk distance (6MWD),\textsuperscript{24,25,44,45} peak oxygen consumption and functional capacity\textsuperscript{24,43,46,47,49} The improvements in these clinical parameters have often been correlated with pulmonary hemodynamic improvement post-PEA.

REFERENCES


